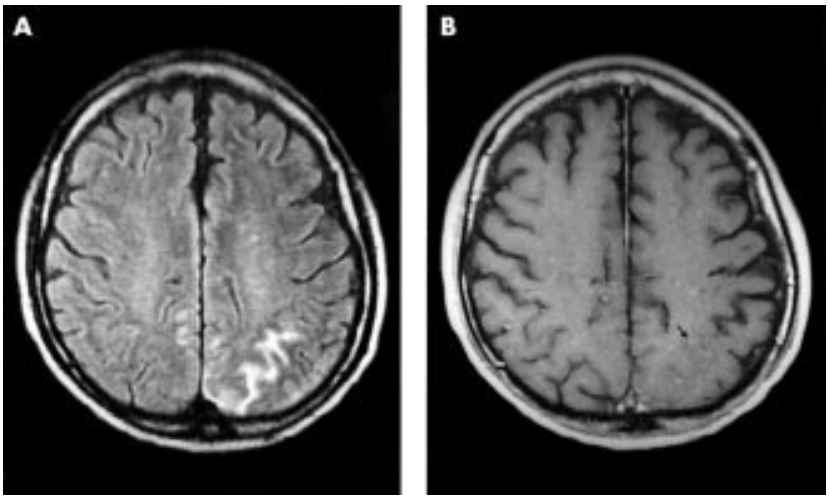

Figure 1 (A) Axial FLAIR (TR 9000/TE 110/TI 2500 ms) section through the centrum semiovale above the level of the lateral ventricles shows abnormal high signal within the left parietal sulci, without intracerebral vasogenic oedema. (B) Corresponding axial enhanced T1 weighted section (TR 540/TE 12 ms) showing obliteration of the normally dark CSF containing sulci (compare with frontal sulci) and subtle enhancement (indicated by arrows).

An 81 year old man had a one week history of progressive lower limb weakness and numbness associated with pain radiating down his right leg. There was no preceding history of infection or trauma. He had no significant past medical history.

On examination, he was alert and rational. There was no slurring of speech or paresis of the extraocular muscles. Cranial nerve and visual field examination was unremarkable and the neck was supple. He had mild proximal symmetrical upper limb weakness (MRC grade 4/5) and bilateral lower limb weakness (MRC grade 3/5). He had difficulty in walking unaided and in tandem walking. Sensory loss to touch was elicited in the distal lower extremities in stocking distribution. Reflexes in all four limbs were absent. The clinical features were consistent with the Guillain–Barré syndrome.

The patient was initially referred to an orthopaedic surgeon for possible lumbosacral spondylitic disease. Unenhanced MRI scans of the lumbar and thoracic spine showed mild degenerative changes and excluded intrinsic cord abnormalities or external compression. Subsequent enhanced cervical spine MRI scans were also negative. He was then referred for a neurological opinion.

Nerve conduction studies revealed significantly prolonged distal motor latency (median motor distal latency 6.7 to 9.8 ms; posterior tibial distal latency 7.7 to 9.4 ms) and reduced conduction velocities in the median (40.3 m/s), ulnar (39.6 m/s), and tibial nerves (35.7 m/s). F responses were prolonged (> 37 ms) or absent in all four limbs. Cerebrospinal fluid examination showed absent cells with raised protein of 0.8 g/l, normal glucose concentration, and positive globulin. Bacterial culture and viral studies were negative.

The first MRI of the brain, obtained to exclude a central cause for weakness and gait abnormalities during the first week of admission, showed left parietal and superior occipital sulcal hyperintensities on the fluid attenuated inversion recovery (FLAIR) sequences, in addition to subtle enhancement with contrast administration (fig 1). A repeat MRI one week later showed mild improvement. A third MRI two months after initial presentation showed resolution of the focal abnormalities.

In the related Miller–Fisher syndrome, MRI changes have been documented in the cranial nerves,[3] spinocerebellar tracts, and pons.[4] We postulate that our MRI findings represent a focal manifestation of a wider immunologically mediated reaction within the subarachnoid cerebrospinal fluid bathed space. This focal sulcal reaction probably represents a local concentration of proteinaceous fluid and correlates well with the CSF findings of high protein levels but an absence of cells.[4] These MRI changes resolved with immunoblobulin treatment and clinical recovery. While MRI changes have been reported in the subcortical regions in demyelinating neuropathies, most probably from focal demyelination, sulcal changes have not been described. Serial MRI studies are a sensitive technique for documenting cerebral cortical abnormalities in this condition, even in the asymptomatic setting as demonstrated here.

Y L Lo, M C Wong
Department of Neurology, Singapore General Hospital, Outram Road, 169608 Singapore

L L Chan
Department of Diagnostic Radiology, Singapore General Hospital

P A C Lim
Rehabilitation Medicine, Singapore General Hospital and Baylor College of Medicine, Houston, Texas, USA

Correspondence to: Dr Lo; gnrlyl@sgh.com.sg

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Muscle tissue oxygenation as a functional tool in the follow up of dermatomyositis

Near-infrared spectroscopy (NIRS) is a direct, non-invasive optical method for measuring local oxygenation and haemodynamics in muscle tissue. Although measurement of local oxygenation by NIRS has been used for the diagnosis of metabolic myopathies, the technique has not previously been applied to inflammatory myopathies. Dermatomyositis is a muscle disorder characterised by complement mediated capillary necrosis, resulting in ischaemia and hypoperfusion. We have now employed NIRS to study the effect of corticosteroid treatment on haemodynamics in muscle tissue in dermatomyositis.

The pathological features of dermatomyositis are characterised by a decreased number of capillaries per muscle fibre and necrosis of single muscle fibres or clusters of fibres at the periphery of the fasciculi.[1] Muscle fibre regeneration and an increased number of capillaries have been shown in dermatomyositis after intravenous immune globulin treatment,[2] but corticosteroids are still considered to be the first line of therapy. In the clinical setting, the effect of treatment is mainly assessed by muscle strength and creatine kinase (CK) levels. Direct measurement of capillary and muscle fibre status can only be done by repeated muscle biopsies. However, apart from the fact that muscle biopsies are invasive, they are also a static representation of muscle tissue at a fixed time point and at a particular location (selection bias).

This is the first time that NIRS, a non-invasive optical method for the measurement of oxygenation and haemodynamics in muscle tissue, has been used to study the effect of treatment in a patient fulfilling the clinical and histological criteria of definite dermatomyositis.[3] A young woman from Aruba, aged 24 years, presented with subacute erythema of the facial skin and severe proximal muscle weakness (arm muscles: mean Medical Research Council (MRC) grade 3; leg muscles: MRC grade 2). Serum CK levels were slightly increased (220 IU/l). Five weeks after the onset of symptoms, treatment with corticosteroids was started in our department at a dose of 60 mg/day (for six weeks), the dose being subsequently tapered. CK levels decreased and muscle strength increased (arm muscles: MRC grade 4; leg muscles: MRC grade 3) in week 12.

Tissue oxygenation was measured by NIRS immediately before treatment was begun and again after three and seven weeks of treatment. NIRS is based on the relative tissue transparency to light in the near-infrared region, and on the oxygen dependent absorption changes of haemoglobin and myoglobin. Using a modification of the Lambert–Beer law, in which physical path length is incorporated to account for light scattering, it is possible to calculate quantitative values for oxygen consumption and blood flow in skeletal muscle. NIRS is non-invasive and measures oxygenation directly in the muscle. Moreover, it is relatively inexpensive, easy to apply, and applicable at the bedside.

In this study, NIRS measurements were obtained using a continuous wave near-infrared spectrophotometer (Oxymon, Biomedical Engineering Department, University

of Nijmegen, Netherlands). Using this spectrophotometer, which generates light at 905, 850, and 770 nm, it is possible to differentiate between oxyhaemoglobin/myoglobin (O_2Hb/O_2Mb) and deoxyhaemoglobin/myoglobin (HHb/HMb). The optical fibres were placed on top of the flexor digitorum superficialis muscle in the same location for all the measurements. Data were sampled at 10 Hz.

Quantitative NIRS values for oxygen consumption ($m\dot{V}O_2$) were calculated by evaluating the rate of decrease in [O_2Hb] during arterial occlusion, as previously described.[4] Reoxygenation rate (ΔO_2Hb) was determined as the rate of initial increase in O_2Hb measured over three seconds immediately after cessation of arterial occlusion. Both $m\dot{V}O_2$ and ΔO_2Hb were calculated at rest and following rhythmic isometric handgrip exercise at various work intensities. Each exercise session consisted of one minute of exercise at a contraction rate of 30/min (50% duty cycle), immediately followed by 45 seconds of arterial occlusion for the calculation of $m\dot{V}O_2$ and ΔO_2Hb. Whereas $m\dot{V}O_2$ is a measure of mitochondrial function at a certain work insentity and is dependent on the vascular capacity of oxygen delivery, ΔO_2Hb reflects the initial recovery rate at which deoxygenated haemoglobin/myoglobin are resaturated.[5] It is therefore directly related to microvascular function. All measurements were performed at the same absolute work intensities.

Figure 1 shows the effect of corticosteroid treatment, measured non-invasively and with relative ease by NIRS, in the patient with severe dermatomyositis. Before treatment was begun, resting $m\dot{V}O_2$ was slightly higher than in healthy controls (0.19 *v* 0.14 ml O_2/min/100 g, respectively). However, $m\dot{V}O_2$ during exercise was about 60% lower than in the controls over the whole range of exercise intensities (fig 1A). After three weeks of treatment, $m\dot{V}O_2$ had already markedly increased. After seven weeks, $m\dot{V}O_2$ had increased even further and was now only 25% below that of the controls, and within the normal range at several work intensities. Serum CK levels were normalised, while muscle strength had increased. ΔO_2Hb (fig 1B) showed similar results, with slow recovery rates before treatment was begun and an increase over all work intensities at the three week and seven week examinations. ΔO_2Hb after seven weeks of treatment exceeded the normal mean value.

As NIRS measures local oxygenation and haemodynamics within the muscle, it can give direct insight into the working microvascular system. ΔO_2Hb increased during treatment, indicating an increase in capillary function. As a result of the increased capillary function and a possible regeneration of muscle fibres, muscular oxygen availability increased, enhancing oxidative capacity—as reflected by the increase in local muscle oxygen consumption.

Although a muscle biopsy will remain indispensable for the diagnosis of dermatomyositis, NIRS is an interesting and non-invasive tool for monitoring the effect of treatment non-invasively and with relative ease. While both serum CK levels and muscle strength are indirect measures, and muscle biopsies provide only a static fingerprint of the muscle, NIRS measures local microvascular and mitochondrial function directly in the intact and working physiological setting.

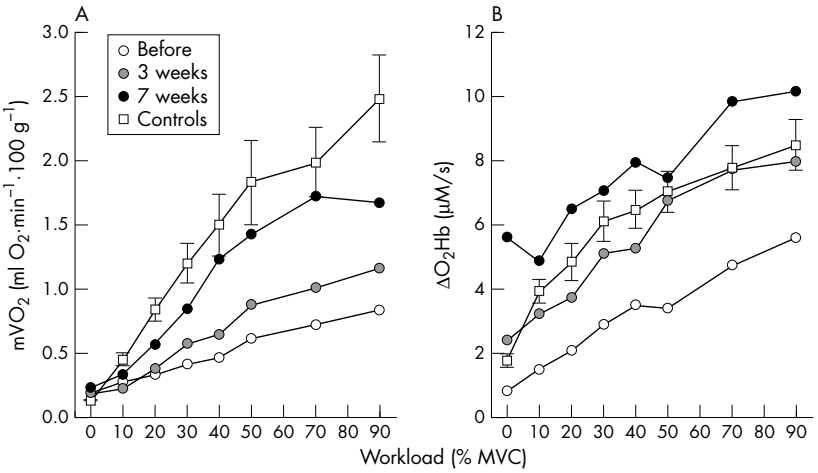

Figure 1 Effect of treatment in a patient with severe dermatomyositis. (A) Muscle oxygen consumption ($m\dot{V}O_2$), and (B) reoxygenation rate (ΔO_2Hb) measured non-invasively by near-infrared spectroscopy at rest and after exercise at different levels of maximum voluntary contraction force (MVC). Mean values ± SD are shown for the controls.

M C P van Beekvelt, R A Wevers, B G M van Engelen
Neuromuscular Centre, Institute of Neurology, University Medical Centre Nijmegen, PO Box 9101, 6500 HB Nijmegen, Netherlands

W N J M Colier
Department of Physiology, University Medical Centre, Nijmegen

Correspondence to: Dr B G M van Engelen; b.vanEngelen@czzoneu.azn.nl

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Propofol in myoclonus status epilepticus in comatose patients following cardiac resuscitation

Myoclonus status epilepticus has been identified as a poor prognosticating sign in comatose patients following cardiopulmonary resuscitation.[1] These vigorous generalised jerks are considered to be the penultimate phenomenon in a severly damaged brain that is difficult to manage and that may cause difficulty in ventilating the patient. Antiepileptic drugs such as phenytoin or benzodiazepines have not been very successful. When the jerks are particularly severe, neuromuscular junction blockers have been recommended.[1] I report on two comatose patients with myoclonus status epilepticus. Propofol in a subanaesthetic dose muted these movements considerably.

A 77 year old patient with a prior history of rheumatoid arthritis was resuscitated at home after sudden collapse. The emergency medical service found no pulse. He was defibrillated, and after resuscitation of approximately 70 minutes, pulse and blood pressure returned. In the coronary care unit, he had generalised myoclonus in the face, limbs, and abdomen muscles and the movements were particularly sensitive to touch. His Glasgow coma score was 3 and brainstem reflexes were intact. These rhythmic jerks interfered with mechanical ventilation and caused repetitive bucking of the ventilator. He was treated with fosphenytoin (phenytoin equivalents 20 mg/kg), which subsequently reduced his blood pressure to 80 mm Hg but which quickly returned to a normal level. He was placed on a propofol infusion titrated to a maximal dose of 65 μg/kg/min, and myoclonus disappeared. After treatment for three hours, propofol was discontinued. An electroencephalogram showed a burst suppression pattern. Myoclonic jerks returned and, in addition, constant blinking was noted. The patient did not awaken after discontinuation of propofol on the second day.

A 19 year old boy was found hypothermic (core temperature of 31°C) in the field after a car rollover. He was resuscitated for 30 minutes before heart rate returned. On admission, his Glasgow coma score was 3. Notable signs were constant facial jerking, biting on the endotracheal tube, and sound sensitive myoclonus jerks in all limbs. Propofol in a dose of 35 μg/kg/min significantly muted myoclonus, although occasional myoclonic jerk was noted in both legs. An electroencephalogram showed a burst suppression pattern. Computed tomography showed poor white-grey matter differentiation, indicating early brain oedema. Care was withdrawn after the patient did not recover from coma after discontinuation of propofol.

Control of generalised myoclonus status epilepticus has been difficult and frustrating. I noted that the use of propofol in a fairly low dose muted myoclonus considerably. The typical dose in the intensive care unit is 5 μg/kg/min, which can then be titrated to 50–